Short communication

Sarcoidosis. Debut as orbital pseudotumour

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ABSTRACT

Case report: 78-Year-old female patient being investigated for a left orbital pseudotumour of 17 days onset. She had proptosis, pain, with no limitation of eye movements, peribulbar edema, and upper eyelid ptosis, compatible with an orbital pseudotumour. The MRI showed an upper outer left orbital mass adjacent to the side of eyeball and an increase in soft tissue at the level of lacrimal gland. The histopathology study of an eyelid lesion demonstrated the presence of non-caseating granulomas.

Discussion: A definitive diagnosis of orbital sarcoidosis was made. Oral treatment with corticosteroids and methotrexate achieved gradual control of the disease.

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Sarcoidosis. Comienzo como pseudotumor orbitario

RESUMEN

Caso clínico: Paciente mujer de 78 años remitida para estudio de pseudotumor orbitario izquierdo de 17 días de evolución. Observamos proptosis, dolor sin limitación de los movimientos oculares, edema periorbitario y ptosis palpebral, clínica compatible con pseudotumor orbitario. La resonancia magnética orbitaria evidencia una masa adyacente al globo ocular ocupando la vertiente superexterna y aumento de partes blandas a nivel de la glándula lagrimal. El estudio anatomopatológico de una lesión palpebral demuestra la presencia de granulomas no caseificantes.

Discusión: Se realiza el diagnóstico de sarcoidosis orbitaria definitiva. El tratamiento oral con corticoides y metotrexato logra el control de la enfermedad.

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Introduction

The most frequent causes of orbital inflammation are dysthyroid orbitopathy, lymph proliferative diseases and in third place orbital pseudotumor which accounts for about 8–11% of all orbital tumors. Pseudotumor is defined by idiopathic inflammation derived from an abnormal response of pleomorphic inflammatory cells and reactive fibrosis. Occasionally, histology reveals a range between granulomatous and non-granulomatous inflammation with formation of fibrosis.

Sarcoidosis is an inflammatory disease which for a number of years was suspected to involve the orbital region, giving rise to a condition simulating orbital pseudotumor, first described by Birch-Hirschfield in 1905 as an inflammatory non-neoplastic process involving the orbit.

Sarcoidosis is a self immune disease of unknown cause that involves T-lymphocytes with the formation of non-caseating granulomas and involving multiple organs, including the eye. In some cases, the disease is latent in organs but is not suspected until histology signals the direction.

A clinic case is described below of a difficult to manage patient in which, after the excisional biopsy of a palpebral lesion a new diagnostic of orbital sarcoidosis was established.

Clinic case report

Female, 78, referred by another center with a diagnostic of left orbital pseudotumor with 17 days evolution. Personal history comprised arterial hypertension, hypotensive cardiopathy, dyslipidemia and transient cerebrovascular accident. In what concerns ophthalmological history, the patient referred cataract surgery in both eyes without other diseases. Best corrected visual acuity in the right eye (RE) of 0.9 and 0.7 in the left eye (LE). LE exhibited 24 mm proptosis measured with Hertel exophthalmometer, periorbital edema inducing secondary mechanics, 3 x 2 mm indurated tumor at the level of the upper eyelid (Fig. 1) and moderate conjunctival chemosis. The patient did not refer having fever the days, although she did refer asthenia and dyspnea during efforts.

Treatment was established with oral prednisone at a dose of 1 mg/kg/day, with subsequent progressive reduction which previously was not prescribed, evidencing significant improvement one week later (Fig. 2), although with persistence of tumefaction caused by said orbital tumor. For this reason, subtenon acetonic acid (Trigonal depot, Bristol-Myers Squibb, Anagni, Italy) was administered, observing significant improvement 2 weeks later. MRI revealed the presence of a solid mass of soft parts measuring 15.1 x 7.9 mm in the subprime orbital, superior and external left region, involving the extracanal compartment, muscular extension toward the eyelid elevator and superior rectus, making contact with the sclera and respecting the optic nerve cleavage plane (Fig. 3).

All the above suggested orbital pseudotumor. Lab examinations discarded thyroid disease, with PCR being of 10.1 mg/dl, ECA, ANA, anti-CCP, ENA, Mantoux and quanti-FERON antiviral serology negative. Chest x-ray showed prominent right pulmonary hilum.

Excisional biopsy of the upper palpebral lesion (Fig. 4) reported a finding compatible with non-caseating granulomas (Fig. 5). Thorax computerized axial tomography was requested, which revealed mediastinic and parahilar right adenopathies, with granulomatous-like nodule at the level of the left inferior lobe. Gallium gammagraphy revealed pathological capture in the left lacrimal gland and slight capture in both pulmonary hila.

Treatment was established with methotrexate (Methotrexate Wyeth® 2.5 mg capsules, Wolfhrathausen, Germany) at a dose of 20 mg/week and prednisone 10 mg/day, after 8 months follow-up (Fig. 6).

Discussion

The most frequent expression of sarcoidosis is granulomatous uveitis, which initially presents in approximately 30–60% of patients with said disease.

Sarcoidosis management criteria are well established by the international sarcoidosis working group (FIWOS criteria), which classified groups as possible, probable, assumed and definitive. In the present case, after histological confirmation we found a definitive sarcoidosis. Even though as can be seen thorax x-ray findings were not conclusive, the diagnostic sensitivity and specificity is lower that to that of computerized axial tomography. However, the ocular findings after biopsy required an exhaustive study of the patient, which evidenced the characteristic lesions of the disease.

The definitive ocular sarcoidosis diagnostic was established with the histological finding of non-caseating granulomas.

The presentation of sarcoidosis simulating orbital pseudotumor is infrequent, with few cases described in the literature. Even so, it must be taken into account in the differential diagnostic of inflammatory pseudotumor. In the present case, the presentation of an easy to access

Fig. 1 – Proptosis, hyperophthalmos, conjunctival chemosis and initial periorbital tumefaction in the left eye, all of which suggests orbital pseudotumor.

Fig. 2 – Slight improvement of proptosis, tumefaction and disappearance of conjunctival chemosis after introducing oral prednisone. Subtenon triamcinolone acetonide injection was decided to apply in order to enhance improvement.
Fig. 3 – Magnetic resonance images of both orbits. High Field 1.5 Tesla equipment, Optima MR360.2012 GE®. Images not enhanced: A. pondered axial T2; B. Coronal acquisition STIR; C. pondered sagittal T2. Solid mass of soft parts (white star) located in the supraorbital, superior and left external region, including extracanal compartment involvement, extension to the muscular plane of the eyelid elevator and superior rectus, making contact with sclera and respecting the left optic nerve cleavage plane. The lesion exhibits high signal behavior in STIR acquisition and low signal in enhanced T2.

Fig. 4 – Indurate nodular lesion without adherence to deep layers, approximately 3 × 2 mm at the level of the left eye upper eyelid. Excisional biopsy thereof was carried out.

Fig. 5 – Histological section 40 × (hematoxylin-eosin staining) of the excisional biopsy sample of the upper LE eyelid, showing typical image of non-caseating granulomas made up of multinucleated giant epithelioid cells and small amount of lymphocytes without central necrosis.

Fig. 6 – After prescribed treatment, palpebral secondary mechanical ptosis was recovered, showing periorbital tumefaction and appearance of the physiological upper palpebral sulcus.
nodule for excisional biopsy was very helpful to establish the definitive diagnostic.

In what concerns treatment, there is a rapid response to oral corticoids, which should be the first therapeutic step. However, occasionally periocular injections must be applied for control purposes, without discarding surgery.

In the case presented herein, early diagnostic was crucial for controlling the disease. In addition, low corticoid doses and methotrexate produced efficient results.

Atypical presentation of sarcoidosis in the present patient simulated orbital pseudotumor, which required the authors to reconsider the differential diagnostic. The literature comprises very few orbital sarcoidosis cases similar to that described herein.

**Conflict of interests**

No conflict of interests was declared by the authors.

**REFERENCES**